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Images

Manifestations of hypomelanosis of Ito

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A 6-year-old boy adopted from China at 1 year of age had an unwitnessed fall with brief loss of consciousness. A non-contrast computed tomography of the head showed an incidental large left hemispheric hypodensity. He had history of anal stenosis and a rectal stricture requiring multiple dilations. His dentist had noticed that some of his teeth were missing and others were small. He did not have history of seizures and had achieved his developmental milestones on time. His skin showed multiple hypopigmented linear patches following Blaschko's lines exclusively on the left arm and leg (Fig. 1). These had remained unchanged since adoption. His neurological exam including fundoscopy was normal. Magnetic resonance imaging (MRI) pre and post gadolinium of the brain showed a predominantly white matter abnormality involving most of the left hemisphere with no contrast enhancement and with dilation of perivascular spaces (Fig. 2).

Hypomelanosis of Ito (HI) is a rare neurocutaneous syndrome. Its diagnosis requires early appearance of nonhereditary, hypopigmented patches involving two or more body segments. Up to 90% of HI cases show CNS manifestations including developmental delay, intellectual disability, epilepsy, autism, and hypotonia.[1] Seventy percent of HI patients have musculoskeletal manifestations including scoliosis, chest wall deformity, or finger abnormalities. Retinal hypopigmentation is most the most common ocular manifestation. Dental, renal, cardiovascular, and endocrine malformations can be also present.[2] Treatment is symptomatic targeting the specific clinical manifestations of each patient.

Figure 1 Patient's arm showing hypopigmented streaks following Blaschko's lines.

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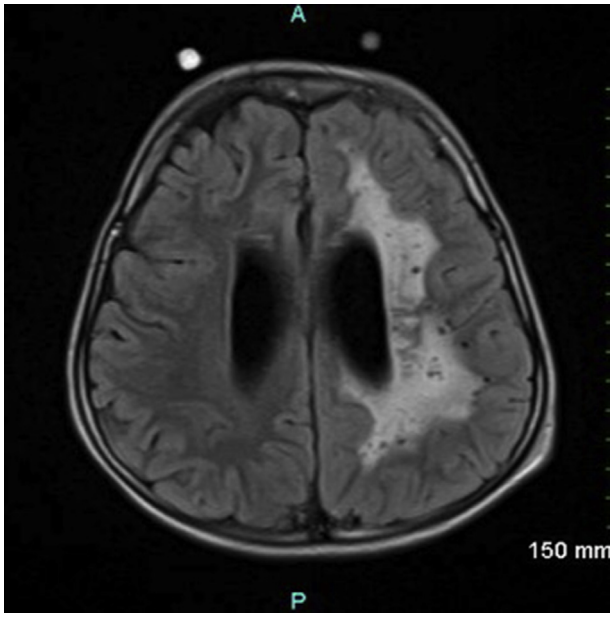

Figure 2 Brain FLAIR MRI shows hyperintense signal predominantly in the white matter of the left hemisphere. There are small cystic dilations of the perivascular space.

Although HI is infrequent, most pediatricians will encounter patients with neurocutaneous syndromes during their careers. Skin findings may be subtle and require a detailed physical examination. Exam with a Wood's lamp may be necessary in children with fair skin. Early recognition of neurocutaneous syndromes by primary care providers can facilitate prompt referral to specialists, close patient follow-up to monitor for complications, and guide genetic counselling.[3]

Conflicts of interest

Drs. Albuja, Shrivastava and Khan have no conflicts of interest to declare in relation to this manuscript.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.pedneo.2018.03.009.